patología
Revista latinoamericana

Patología 2009;47(3):234-8

Case report

A mediastinal gastric cyst as cause of non-specific abdominal pain

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RESUMEN

Paciente del sexo masculino de 17 años de edad, que fue admitido al hospital por padecer dolor abdominal inespecífico e intermitente, de un día de evolución, acompañado de vómitos. Tenía antecedentes de los mismos síntomas durante toda su vida y había sido tratado anteriormente con tres laparotomías, aparentemente por un síndrome de oclusión intestinal, sin encontrar la causa de sus síntomas. Debido a la ausencia de datos clínicos y resultados de laboratorio para determinar una causa específica del dolor, se realizó una tomografía computarizada, que reveló una lesión quística localizada en el mediastino posterior y espina bífida en S1. El tratamiento se llevó a cabo mediante una toracotomía con resección completa de la lesión. El diagnóstico definitivo fue un quiste gástrico mediastinal, con úlceras pépticas crónicas activas, gastritis superficial y gastritis crónica atrófica y folicular. El dolor abdominal inespecífico es un motivo común de ingreso hospitalario y en muchas ocasiones no se demuestra una causa aparente. El quiste gástrico mediastinal puede ser el origen atípico de dolor abdominal inespecífico, lo que hace el diagnóstico difícil para el clínico. El diagnóstico diferencial es amplio, por lo que la sospecha de esta enfermedad se hace mediante estudios de imagenología. El tratamiento de elección es la resección total de la lesión. Sin embargo, hay pocos casos en los que la resección total no puede llevarse a cabo, lo que disminuye la tasa de éxito.
Palabras clave: quiste gástrico mediastinal, dolor abdominal inespecífico, malformaciones quísticas del intestino anterior.

ABSTRACT

A 17-year old male with a 1-day history of non-specific and intermittent abdominal pain, accompanied by vomiting was admitted into the hospital. He had a background of the same symptoms throughout his life, treated with three laparotomies, supposedly due to intestinal occlusion syndrome, without findings that suggested an etiology of his symptoms. Due to the absence of clinical data and laboratory results to determine a specific etiology, a computed tomography scan was performed. The screen revealed a cystic-like lesion located at posterior mediastinum and a hidden *spina bifida* at S1 was discovered. The treatment was a thoracotomy with complete resection of the mass. A gastric mediastinal cyst was the definitive diagnosis. The cyst had chronic active peptic ulcers, superficial gastritis, and atrophic and follicular chronic gastritis. Non-specific abdominal pain is a common cause of hospital admission, and most of the times it has no apparent etiology. Gastric mediastinal cysts may be an atypical cause of non-specific abdominal pain, making the diagnosis difficult for the clinician. The differential diagnosis is broad, therefore in many cases suspicion of this disease is made mainly by imaging studies. The treatment of choice is total surgical excision of the cyst. Unfortunately there are few cases in which total excision can not be done, leading to other surgical approaches that may decrease the rate of success.
Key words: gastric mediastinal cyst, non-specific abdominal pain, cystic foregut malformations.

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Received: March, 2009. Accepted: May, 2009.

This article must be quoted: Iglesias-Miramontes G, Ramos-Martínez E, Hernández-Saldaña R, Ordoñez-Solorio LA. A mediastinal gastric cyst as cause of non-specific abdominal pain. Patología Rev Latinoam 2009;47(3):234-8.
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Abdominal pain is a common cause of admission to the hospital through the emergency department. An exhaustive patient interview and complete physical examination may lead us to order specific complimentary tests, which include laboratory exams, and imaging studies. But, what happens when findings do not guide us to a diagnostic suspicion? Then, the clinician has a real challenge. Can a non-specific abdominal pain episode have an explanation? Can a gastric mediastinal cyst be a cause of non-specific abdominal pain?

Usually, gastric mediastinal cysts are uncommon, produce obstructive symptoms, and sometimes are asymptomatic. But they can also cause unspecific signs and/or symptoms. The suspicion of the disease is made by imaging studies and the diagnosis established after removal of the cyst and histopathological examination. Then, after a clinicopathological correlation, the pain episode can be explained.

CASE REPORT

A 17-year-old male presented to the Emergency Department with a 1-day history of intense abdominal pain in upper quadrants, with irradiation to the dorsum, with no more symptoms. He had previous episodes of similar abdominal manifestations accompanied by vomiting. His supposed diagnosis was an intestinal occlusion syndrome, which led to laparotomies at 6, 10 and 11 years of age, with no specific findings.

On physical examination, his vital signs were normal, intense pain was elicited by deep palpation in the *epigastrium*, right and left *hipochondria*, accompanied by muscle rigidity but no rebound tenderness. No masses where palpated.

Laboratory studies where in the normal parameters. A complete double contrast (oral and intravenous) abdominal computed tomography (CT) scan, performed with a Phillips MultiSlice Brilliance 64 CT, showed in first instance the separation of the right paravertebral line, and a hidden *spina bifida* at S1 (Figure 1). The thoracic window confirmed the separation mentioned above caused by an extrapulmonary, para-aortic and anterolateral thin-walled tumoration of 83 x 39 x 40 mm, and passive atelectasis of the right posterior pulmonary segment (Figure 2A). The abdominal window registered a supradiaphragmatic bilobulated cystic-like lesion, accompanied of an intrinsic hyperdense nodular formation of 18 mm, that had no modifications within the different phases of the study (Figures 2B-D).

During thoracotomy, an irregular-shaped, rough, brown-colored tumor of 7.3 x 4.6 x 3 cm was totally excised (Figure 3A-B). Histopathologically, a 36.2 g cystic malformation of the posterior mediastinum was found, containing gastric *fundus-corpus* and antral mucosa, submucosa, *muscularis propria*, and serosa. The specimen showed superficial gastritis in the *fundus-corpus* mucosa

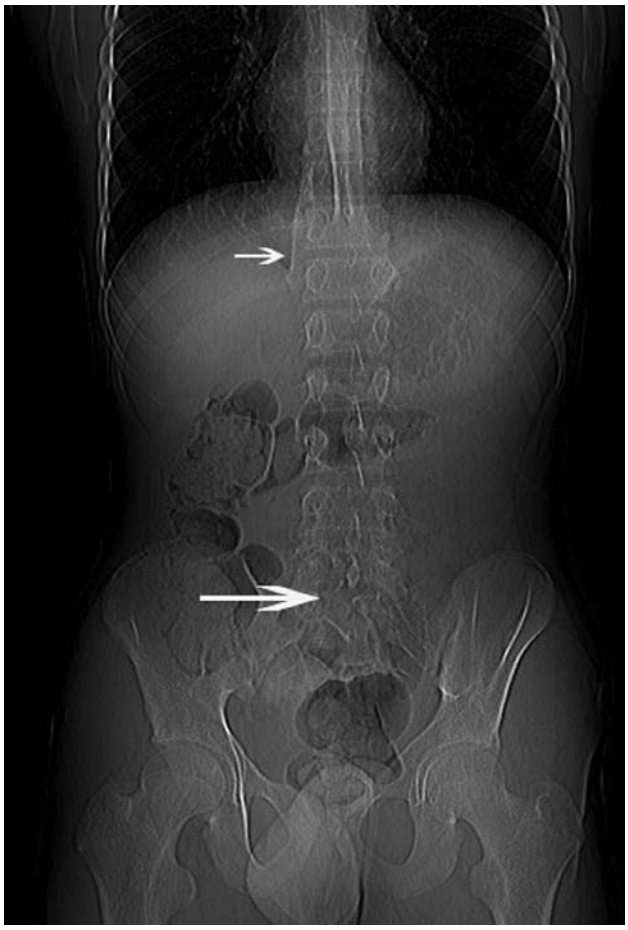

Figure 1. CT survey shows the separation of the right paravertebral line (small arrow), and a hidden *spina bifida* at S1 (large arrow).

(Figure 4A-B), and chronic atrophic and follicular gastritis in the antral mucosa (Figure 5A-B). The mucosa also had chronic active ulcers with eritrocytes (which corresponded to the intracystic nodular-like lesion demonstrated in the CT), neutrophils, edema, granulation tissue, plasmatic cells and eosinophils (Figure 6A-B).

Post-operative evolution was asymptomatic, recovering *ad integrum* in his pulmonary physiologic parameters, and having no more symptoms episodes after 6 months follow up.

DISCUSSION

Mediastinal masses are considered a relatively uncommon group of diseases encountered in thoracic pathology.[1] From these, cysts are benign, uncommon, but an important group

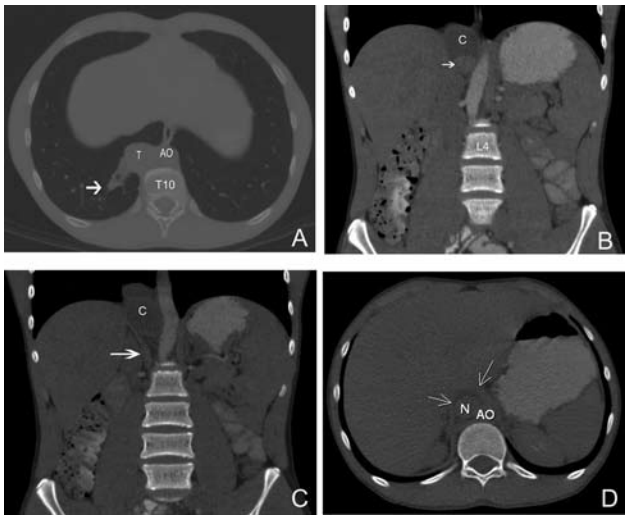

Figure 2. A. CT, axial view, thoracic window at T10 level shows a paraaortic, right anterolateral tumoration (T), with a zone of atelectasis of the posterior pulmonary segment of the right inferior lobe (arrow). **B.** Coronal view, abdominal window, arterial phase at L4 level confirms the presence of a thin-walled cystic tumoration (C) and a nodular-like lesion in its interior (small arrow). **C.** Bilobulated shape of the lesion above the diaphragmatic crura (arrow). **D.** CT, axial view, abdominal window, venous phase at T12 shows the relationship between the intracystic node (N) and the aorta (AO) with the diaphragmatic crura (large arrow). T, tumoration; AO, aorta; C, cystic lesion; N, nodular-like lesion.

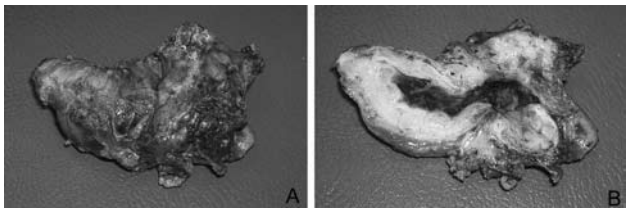

Figure 3. A. Macroscopic aspect of the mediastinal cyst, the external wall has hemorrhagic areas. **B.** Mediastinal cyst that resembles the gastric wall. The mucosa presents ulcerated hemorrhagic zones.

of lesions because their recognition and complete removal prevent serious consequences.[2]

Mediastinal cysts can be classified and defined according to their embryologic origin, epithelial lining and locations as follows: thymic cysts, mesothelial (pericardial and pleural) cysts, cystic hygromas, thoracic duct cysts, and cystic malformations of the foregut.[3]

Cystic malformations of the foregut are as well classified according to their epithelium lining which include respiratory, esophageal, gastric or enteric.[4] Therefore, gastric cysts of the mediastinum belong to the group of cysts that are malformations of the foregut.[3,4]

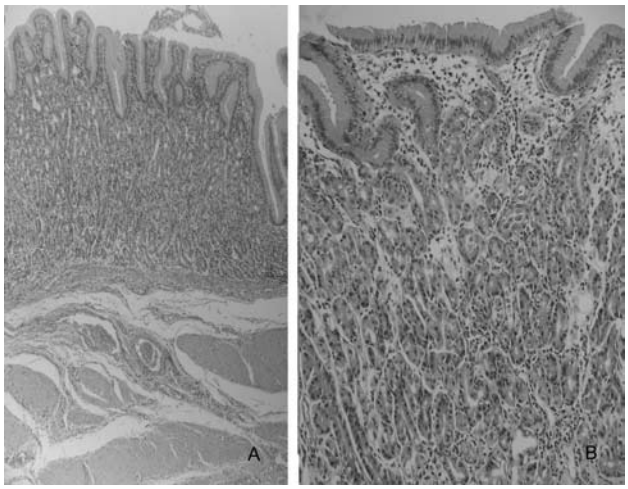

Figure 4. A. Cyst wall with corporal mucosa, submucosa, and *muscularis propria* (H-E x 40). **B.** *Fundus-corpus* mucosa shows superficial gastritis (H-E x 100).

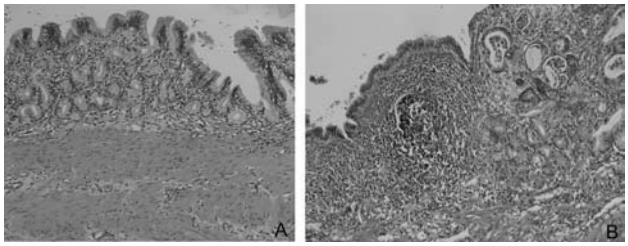

Figure 5. A. Cyst wall with antral mucosa, a small area with submucosa, and *muscularis propia* (H-E x40). **B.** Antral mucosa presents chronic follicular gastritis (H-E x 100).

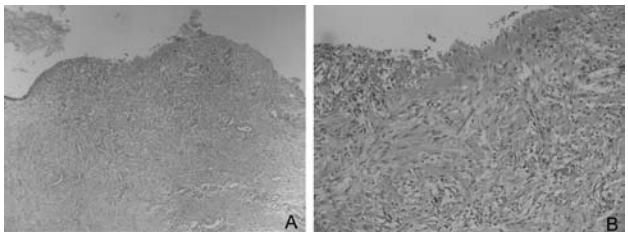

Figure 6. A. Chronic active peptic ulcer. At the upper right corner, the superficial epithelium is preserved (H-E x 40). **B.** Detail of the ulcer described previously (H-E x 100).

From all mediastinal masses, benign cysts account about 18.4%, from which 42% are cystic malformations of the foregut.[1] Gastric cysts correspond to just 1% of all cystic malformations which is clear evidence of the peculiarity of this intrathoracic pathology.[1]

Clinical features include disphagia, cough, pyrexia, hematemesis, weight loss or poor weight gain, retrosternal chest pain, and vomiting; or the patient can be asymp-

tomatic.[1,4,5] Gastric mediastinal cysts are proposed to be considered as a "distinct clinopathological entity" because of "their propensity to produce symptoms in early life", a fatal outcome if undiagnosed, an excellent outcome after complete surgical remove.[4] We agree with this last observation due to the fact that in this particular case, this lesion began symptoms in our patient in early life with a misdiagnosed occlusion syndrome, probably because of persisting vomiting, furthermore he had three different episodes of these symptoms and they lead to an incorrect diagnostic and therapeutic approach, but at an age in which he could explain and describe his symptoms, non-specific abdominal pain comes as a new clinical feature that has not been reported before.

Non-specific abdominal pain is defined by exclusion as an abdominal pain episode that lasts no more than seven days in which no cause could be found.[6] During childhood, it is a common reason of urgent admission to the hospital.[7] There have been some clinical entities implied in non-specific abdominal pain such as viral enteritis, *Streptococcus* sp infection, abdominal aortic aneurysm, mesenteric lymphadenitis, intestinal obstruction, midcycle pain (*mittelschmerz),* menstrual pain, and irritable bowel syndrome.[7] Some studies attempted to imply anxiety and depression as psychological factors related to non-specific abdominal pain episodes, but it could not be proved at all.[6]

Most of mediastinal foregut malformations are accompanied by vertebral malformations such as scoliosis, *spina bifida*, hemivertebrae and incomplete fusion of the neural arches,[8] and gastric cysts are not the exception. In this case, our patient has a hidden *spina bifida* at S1. Among all theories that have tried to explain the origin of these cysts, the most accepted now-a-days was proposed by Veeneklass,[8] who made an important observation due to the fact that these lesions are accompanied by vertebral malformations, especially the upper thoracic vertebrae. He wrote: "I should like to center the interest on the spine". When the notochord begins to develop, approximately at the 18-day aged embryo, there is a migration of ectodermal cells from the primitive pit to the primitive streak, and as it forms, it fuses with endodermal cells, which degenerate later, and the ectoderm tissue is open to the yolk sac. Then, the notochordal plate folds inward to form the notochord.[3,9] The vertebral bodies will form around the notochord.[3] By an unknown cause, some endoderm might fail to degenerate or separate from the notochord, remaining in connection leading to the formation of the foregut malformations.[3,8] By some reason, the specific level or localization of the vertebral anomalies have not been documented, perhaps most of them are close to the mediastinal gastric cyst, as noted by Veneeklass.[8]

However, in this case our patient has a vertebral malformation caudally far away from the localization of the primary mediastinal lesion, whereas normal thoracic and lumbar vertebrae are present. Here it comes that cystic malformations of the foregut are part of the split notho-cord syndrome.[10] Part of the ectoderm from which the nothocord then develops are split or displaced to either one side so that two notochordal centers form, this is the explanation of congenital scoliosis.[10] If we try to complement these concepts, then probably the alteration of the notochord could be along all the notochordal plate and not just only in a particular site where the cysts and the vertebral anomaly are found, now explaining why a complete vertebral anomaly such as scoliosis, and far away vertebral anomalies, such as first vertebrae of the *sacrum*, have been reported.

The gastric cysts are spherical masses with a rough wall.[2] The thickness of the wall varies from 1 to 6 mm;[2,8] the lining epithelium is gastric mucosa, with gastric glands and cells –such as chief and parietal cells–; a secreting-mucosa may be present. Renin, pepsin and hydrochloric acid are reported in the analysis of the secreting fluid.[11] There can be present other type of epithelium in the same cyst such as small-intestinal, duodenal, respiratory, and esophageal.[11] *Muscularis mucosae* is present resembling that of stomach, including Auerbach's plexus' cells and ganglia. A two, and sometimes three, layers of *muscularis propria* may be present. Serosa is sometimes reported, as in this case.

Complications include airway obstruction,[1] and those "produced by peptic digestion",[4] such as pneumonia, empyema, pulmonary hemorrhage, osteomyelitis, and persistent sinuses of the chest wall and neck.[4]

The diagnosis is made with the support of imaging studies. Endoscopic ultrasound has been reported as a minimal invasion alternative of diagnostic approach, even with a better accuracy than CT and magnetic resonance.[12] Fine needle aspiration guided by endoscopic ultrasound can be done for diagnostic purposes, but with a questionable risk-benefit due to the risk of developing mediastinitis and infection.[12]

Treatment of mediastinal cysts is surgical with complete surgical resection of the cyst.[1] Gastric cysts are not the exception. It can be done by thoracothomy or by thoracoscopic surgery.[1,13] Some reports have shown an advantage of minimal invasion surgery in children against classic procedure.[13,14] Certain cyst morphology and locations, such as a common wall with the bronchus and/or esophagus, or a subcarinal position, may lead to thoracotomy as a preferable procedure because it offers less technical difficulty and risk of surgical complications during the dissection.[13] Usually, after a complete removal of the cyst, the outcome is excellent.[1,4]

CONCLUSIONS

Mediastinal gastric cysts are uncommon among thoracic pathology. Suspicion of this disease is difficult based only on clinical findings. They may be asymptomatic, but symptoms, when present, are related to the location, size, and presence of ulceration, ischemia or perforation. In this case, abdominal obstructive symptoms were present since early life; however, our patient had been misdiagnosed through his life leading to three unnecessary abdominal surgeries. Now-a-days, he can explain what he feels, and that was abdominal pain. This can be explained by the fact that cystic foregut malformations develop almost all the structures implied in the embryogenesis and organogenesis of a normal organ, including nerves. So, the perception of abdominal pain should be because of the visceral innervations that develop within the lesion, presenting positive symptoms when an active injury, such as an acute ulcer or hemorrhage, inside the lesion was present, as it was noted in this case. Imaging studies such as ultrasound, endoscopic ultrasound, fine needle aspiration guided by endoscopic ultrasound, computed tomography, and magnetic resonance may offer the first clue for suspicion of the disease. Most of the cases are accompanied by vertebral malformation close to them, most frequently seen at cervical and thoracic vertebrae. In this case, there is a vertebral malformation caudal to the mediastinal lesion, not even closer the lesion, and could be explained by unifying and conciliating theories about the explanation of this malformation which includes the split notochord syndrome theory. A non-specific abdominal pain may have an attributable cause in few cases. The treatment is always surgical with a complete removal of the mass, either by thoracotomy or thoracoscopic approach, leading to the definitive diagnosis after anatomopathological study, and clinicopathological correlation. The outcome is excellent if there are no complications.

Acknowledgements

We want to thank the Department of Radiology of Hospital CIMA Chihuahua for their contributions to this case, especially to Dr. Jesús Alberto Beltrán Marín who well-documented this case, and made a precise radiologic diagnosis, which permitted us to offer a correct approach to the treatment of the patient.

REFERENCES

1. Wychulis AR, Payne WS, Clagett OT, et al. Surgical treatment of mediastinal tumors. A 40 year experience. J Thorac Cardiovasc Surg 1971; 62:379-85.
2. Abell MR. Mediastinal cysts. Arch Pathol 1956; 61:360-79.
3. Allen MS, Payne WS. Cystic foregut malformations in the mediastinum. Chest Surg Clin North Am 1992;2:89-104.
4. Chitale AR. Gastric cysts of the mediastinum-A distinct clinopathological entity. J Pediatrics 1969;75:104-10.
5. Sirivellla S, Ford WB, Zikria EA, et al. Foregut cysts of the mediastinum-Results in 20 consecutive surgically cases. J Thorac Cardiovasc Surg 1985;90:776-89.
6. Raheja SK, McDonald PJ, Taylor I. Non-specific abdominal pain-an expensive mystery. J R Soc Med 1990;83:10-11.
7. Gray DWR, Collin J. Non-specific abdominal pain as a cause of acute admission to hospital. Br J Surg 1982;74:239-42.
8. Veeneklass GHM. Pathogenesis of intrathoracic gastrogenic cysts. Am J Dis Child 1952;83:500-7.
9. Moore KL, Persaud TVN. Embriología Clínica. 6ª ed. México: McGraw-Hill Interamericana, 2002;pp:68-75.
10. Le Roux BT. Intrathoracic duplication of the foregut. Thorax 1962;17:357-362.
11. Salyer DC, Salyer WR, Eggleston JC. Benign developmental cysts of the mediastinum. Arch Pathol Lab Med 1997;101:136-9.
12. Wildi SM, Hoda RS, Fickling W, et al. Diagnosis of benign cyst of the mediastinum: the role and risks of EUS and FNA. Gastrointest Endosc 2003;58:362-8.
13. Michel JL, Revillon Y, Montupet P, et al. Thoracoscopic treatment of mediastinal cysts in children. J Pediatr Surg 1998;33:1745-8.
14. Bratu I, Laberge JM, Flageole H, et al. Foregut duplications: is there an advantage to thoracoscopic resection? J Pediatr Surg 2005;40:138-41.